Disorder of the facial nerve

Embryology

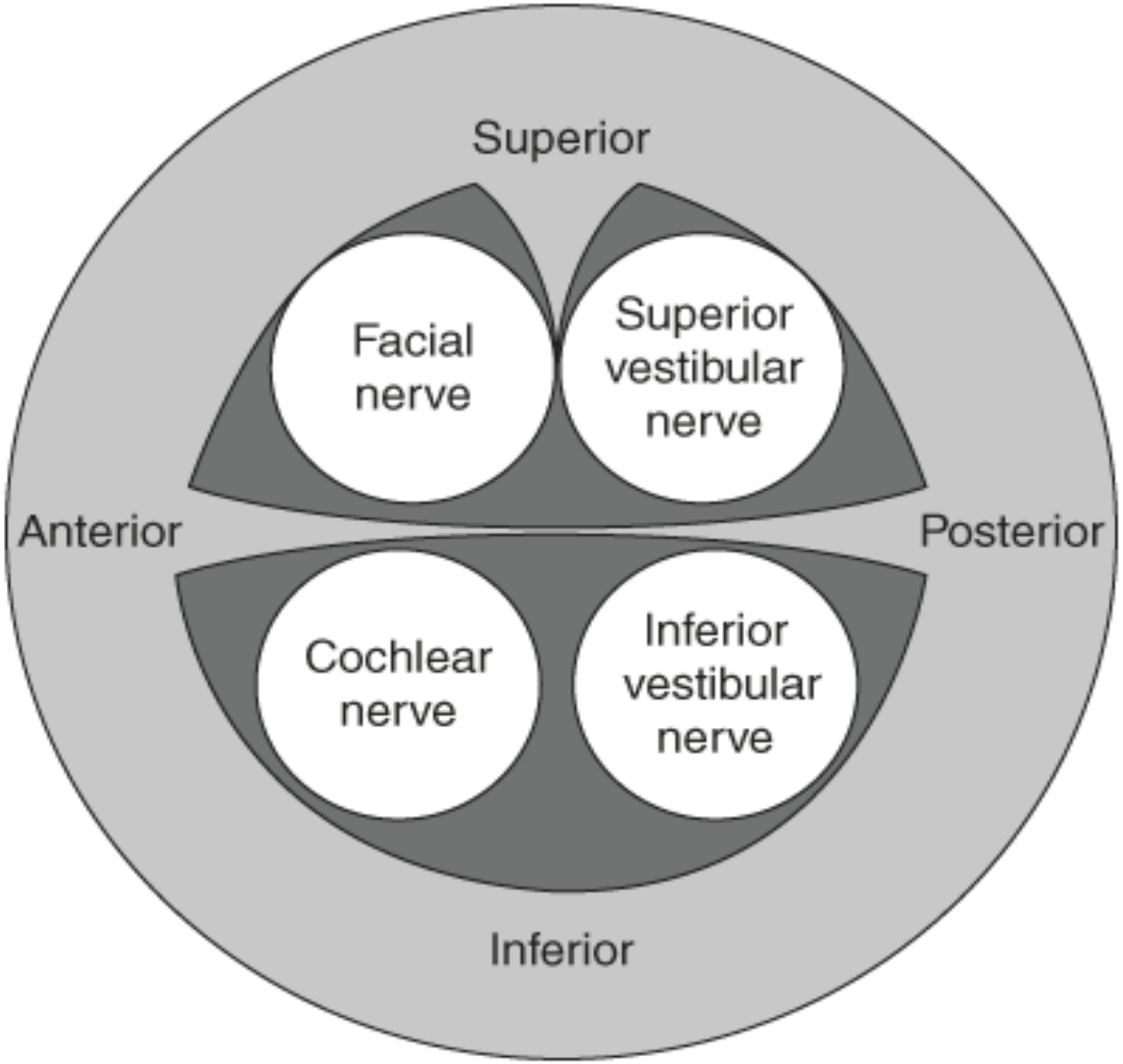
The main pattern of the nerve complex course , branching pattern and the relationship is established during the first 3 months of gestation . during this period the muscles of facial expression also differentiated , became functional and actively contracted and the nerve is not full developed till the 4 years of age

The facial n. develop with in the 2nd pharyngeal arch at the same time of the external and middle ear developed (1st arch) so abnormality of the facial n. should be anticipated whenever there is associated malformation in the external or the middle ear

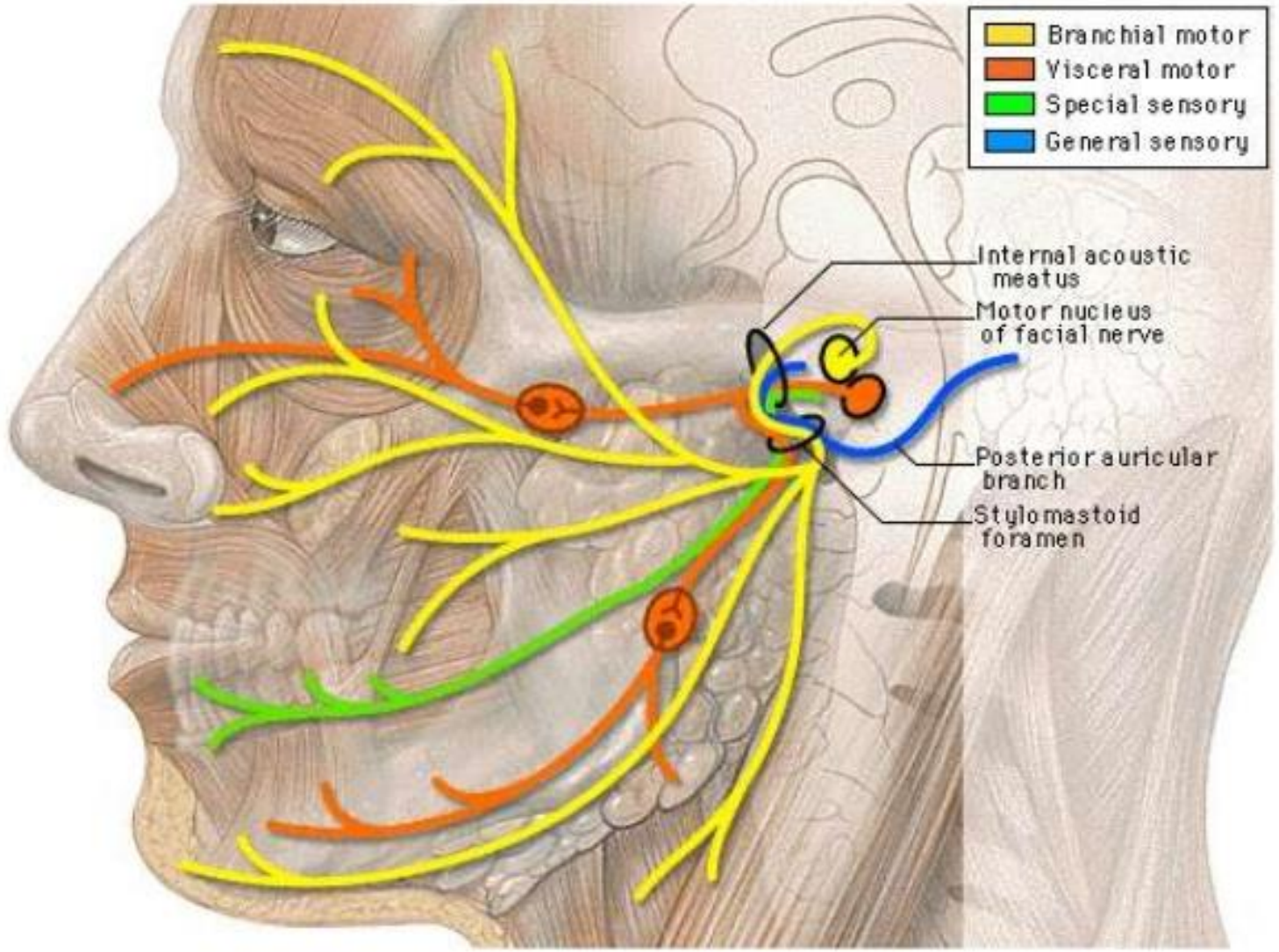
Anatomy

1. Nucleus lies deeply within the substance of the pons is that situation it is closely related to the V nucleus
2. Fibers travel a circuitous route at the first backward to encircle the V1 CN nucleus in the floor of the 4th ventricle and then forward through the pons to emerge on its surface then anterolateral to enter the petrous temporal bone
3. With in the cranial cavity it is closely related to the Viii CN

4. In the intrapetrous part , the facial n. and its sensory root accompany the Viii CN in the internal auditory canal her it has anastamotic with the vestibular nerve at the bottom of this canal , it enter the facial canal which at first runs laterally above the vestibule of the labyrinth until it turns backward through a right angle on the medial wall of the promontory and then fenestera vestibuli


Superior
Facial nerve
Superior vestibular nerve
Anterior
Posterior
Cochlear nerve
Inferior vestibular nerve
Inferior

At the medial wall of the aditus it curves downward to emerge on the inferior surface of temporal bone at the stylomastoid foramen to run forward within the parotid gland
The N. supplies the following
1.Motor : muscles of facial expression , stapedius muscle , posterior belly of diagastric muscle , and stylohyoid muscle
2.Sensory : to the concha and to the parts behind the auricles
3.Autonmic fibers : for lacrimal , submandibular and sublingual glands together with glands at the nasal and oral cavities (secretomotor and vasodilator)
4.Speacial nerve taste via chorda tymapni branch to supply anterior 2/3 of the tongue

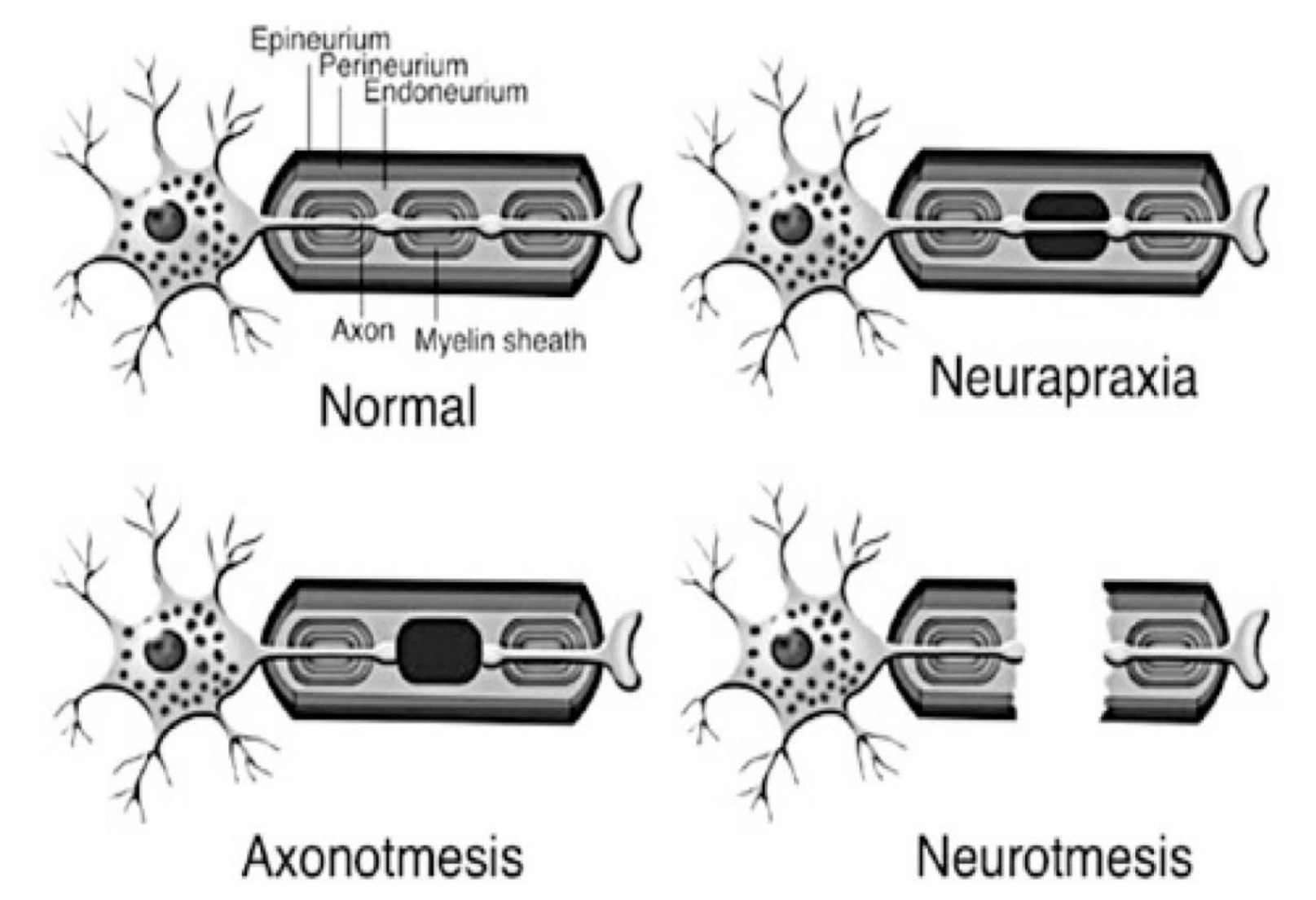

Branchial motor
Visceral motor
Special sensory
General sensory
Internal acoustic meatus
Motor nucleus of facial nerve
Posterior auricular branch
Stylomastoid foramen

Types of nerve injury

1. **Neurapraxia** is defined as a reversible blockage of the transmission of nerve impulses due to pressure on the nerve fibers. Release of the pressure usually results in rapid and complete recovery of the function with no distal Wallerian degeneration

2. **Axonotmesis** is a more severe .injury and involves the blockage of axoplasmic flow Although endoneural tubules are preserved, distalWallerian degeneration occurs

3. **Neurotmesis** total N. transaction


Epineurium
Perineurium
Endoneurium
Axon
Myelin sheath
Normal
Neurapraxia
Axonotmesis
Neurotmesis

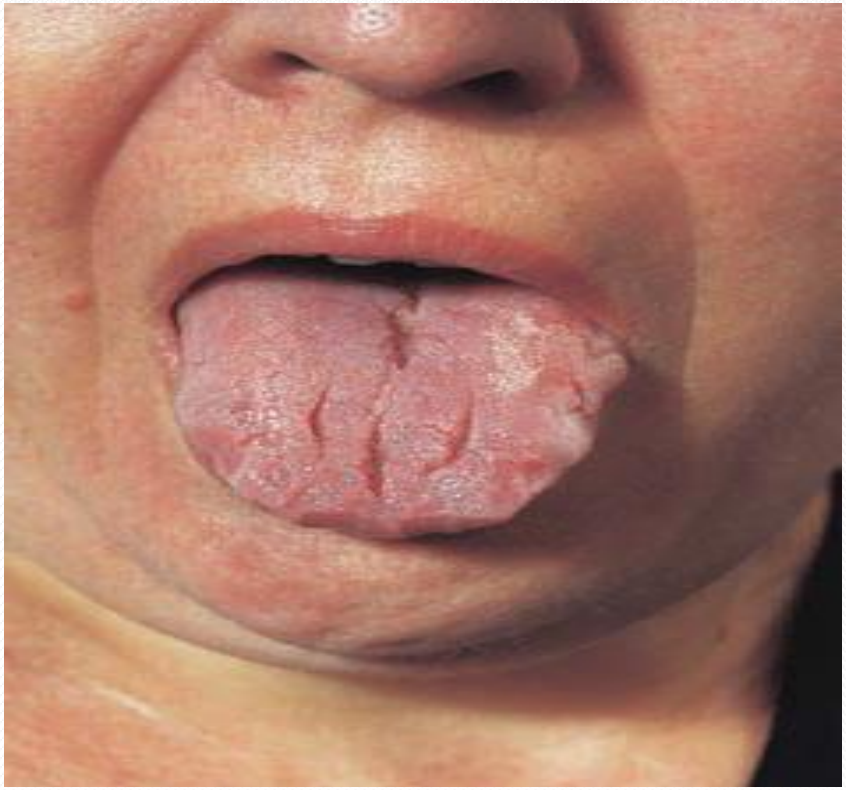
the history of the onset of palsy, whether complete or incomplete, sudden or progressive progressive facial nerve palsy over a period of more than three weeks, or an incomplete facial nerve palsy that does not start to recover after three to six weeks, should make the clinician suspect an underlying neoplasm as the cause and should dictate

the need for further investigations

Ipsilateral recurrent facial nerve palsy can happen in idiopathic palsy, Melkersson–Rosenthal syndrome and tumours

In Bell's palsy recurrence is 13 % and family history is 2.5 times more

Melkersson-Rosenthal syndrome, a condition also characterized by alternating recurrent facial nerve palsy associated with facial oedema, fissured tongue and a positive family history



is almost always benign

bilateral concurrent facial nerve paralysis is most probably associated with a

systemic condition, such as Guillain–Barre´ syndrome (most common), leukaemia, sarcoidosis, Lyme disease, rabies, infectious mononucleosis

physical examination

thorough head, neck, otological and cranial nerve examination is the absolute minimum required when evaluating facial nerve dysfunction

complete or incomplete facial nerve palsy

localize the lesion intracranial , intratemporal or textratemporal

facial nerve palsy may be the first presentation of systemic illness

If symptoms or signs of other cranial nerves deficits are present, a central or systemic cause should be suspected.

Sparing of forehead movement is considered to be characteristic of a central lesion. However, it should be remembered that normal movement can also be seen in facial nucleus lesions and peripheral lesions of the temporal

House–Brackmann system. It has become the most widely used scheme and has been endorsed by the American Academy of Otolaryngology – Head and Neck Surgery. In the House–Brackmann system, grade I is normal function, grade VI is complete absence of facial motor function and grades II–V are intermediate

Table 241c.2 House-Brackmann staging system.

Degree of injury	Grade	Definition
Normal (1°)	I	Normal symmetrical function in all areas
Mild dysfunction (barely noticable) (1–2°)	II	Slight weakness noticeable only on close inspection Complete eye closure with minimum effort Slight asymmetry of smile with maximal effort Synkinesis barely noticeable, contracture or spasm absent
Moderate dysfunction (obvious difference) (2–3°)	III	Obvious weakness, but not disfiguring May not be able to lift eyebrow Complete eye closure and strong but asymmetric mouth movement with maximal effort Obvious, but not disfiguring synkinesis, mass movement or spasm
Moderately severe dysfunction) (3°)	IV	Obvious disfiguring weakness Inability to lift eyebrow Incomplete eye closure and asymmetry of the mouth with maximal effort Severe synkinesis, mass movement, spasm
Severe dysfunction (3–4°)	V	Motion barely perceptible Incomplete eye closure, slight movement corner mouth Synkinesis, contracture and spasm usually absent
Total paralysis	VI	No movement, loss of tone, no synkinesis, contracture or spasm

Special investigations

There are 3 important issue when confronted with facial n palsy

The cause

The site of lesion

The prognosis

TOPODIAGNOSTIC TESTING

These test aim to localize the site but has no prognostic value

Table 241c.3 Topodiagnostic tests.

Test	Nerve branch assessed	Technique considerations	Assessment/Outcome
Schirmer test (**Figure 241c.5**)	Greater superficial petrosal nerve	Strips of paper are placed in the inferior conjunctival fornix for five minutes and the length of paper moistened is compared between eyes	>75% unilateral decrease in lacrimation, or a bilateral decrease in lacrimation (less than 10 mm for both sides at five minutes)
Stapedial reflex	Nerve to stapedius muscle	See Chapter 232, Psychoacoustic audiometry	Present or absent
Electrogustometry	Chorda tympani	The tongue is stimulated electrically to produce a metallic taste and the two sides are compared	Threshold of the test is compared between sides
Salivary flow testing	Chorda tympani	Warthin's ducts are cannulated and salivary flow is measured over time following a gustatory stimulus (6% citric acid on anterior part of tongue)	A reduction of 25% is considered abnormal

Used mainly in complete facial n palsy not in incomplete facial n palsy

Currently, the two most helpful are the ENoG and EMG

Electroneuronography (ENoG)

Consider the most valuable prognostic indicators among electrophysiological test and the main indication is acute onset complete facial paralysis

Electromyography (EMG)

Electromyography records active motor unit potentials of the orbicularis oculi and orbicularis oris muscles during rest and voluntary contraction.

EMG can be used to determine:

_ if a nerve in question is in fact in continuity

_ if there is evidence of Wallerian degeneration

_ if there are early signs of reinnervation

Intraoperative nerve monitoring

Intraoperative monitoring includes continuous EMG measurement from peripheral facial muscle groups and electrical stimulation of the facial nerve itself or its branches to obtain a CMAP

it has a place in cerebellopontine angle (CPA) tumour surgery, in revision mastoid and parotid surgery, and in surgery of congenital ear abnormalities. Other issues to consider are medicolegal issues

Ct scan

The tympanic portion is probably easiest to identify on axial computed tomography (CT) scans at the level of the body of incus and its short process. From there on, it can be followed proximally and distally towards the labyrinthine and descending parts, respectively

The descending or mastoid segment is best visualized in coronal or saggital views.

MRI

Owing to the rich perineural arteriovenous plexus which surrounds the facial nerve, enhancement may be observed normally on T1-weighted magnetic resonance imaging (MRI) with the use of contrast agents. It is usually observed in more than one segment, more commonly in the geniculate ganglion and the tympanic segments and it may enhance asymmetrically between

Causes of facial palsy

Birth

Moulding

Forceps delivery

Dystrophia myotonica

Moebius syndrome (facial diplegia associated with other cranial nerve defects)

Trauma

Basal skull fracture

Facial injuries

Penetrating injury to middle ear

Altitude paralysis (barotauma)

Scuba diving (barotauma)

Lightning

Neurological

Opercular syndrome (cortical lesion in facial motor area)

Infection

Otitis externa
Otitis media
Mastoiditis
Chicken pox
Herpes zoster cephalicus (Ramsay Hunt syndrome)
Encephalitis
Poliomyelitis (type I)
Mumps
Infectious mononucleosis (glandular fever)
Leprosy
Coxsackie virus
Malaria
Syphilis
Scleroma
Tuberculosis
Botulism
Acute haemorrhagic conjunctivitis (enterovirus

Mucormycosis
Lyme disease

Metabolic

Diabetes mellitus

Hyperthyroidism

Pregnancy

Hypertension

Acute porphyria

Neoplastic

Cholesteatoma

VIIth nerve tumour

Glomus jugulare tumour

Leukaemia

Meningioma

Haemangioblastoma

Sarcoma

Carcinoma

Anomalous sigmoid sinus

Haemangioma of tympanum

Facial nerve tumour

Schwannoma

Teratoma

Toxic

Tetanus
Diptheria
Carbon monoxide
Iatrogenic
Mandibular block anaesthesia
Anti-tetanus serum
Vaccine treatment for rabies
Post-immunization
Parotid surgery
Mastoid surgery
Embolization
Dental
Idiopathic
Bell’s, familial
Melkersson–Rosenthal syndrome (recurrent alternating facial palsy, furrowed tongue, faciolabial oedema)
Temporal arteritis
Multiple sclerosis
Myasthenia gravis

Idiopathic (Bell's) palsy

Bell's palsy means facial paralysis that has signs and symptoms consistent with the disease and no cause was found

It includes paralysis of paresis of all muscle groups on one side of the face; sudden onset; absence of signs of central nervous system disease; and absence of signs of ear or CPA disease

Male to female ratio is equal

Recurrence rate 4.5 – 15 %

4.1 % has family history

The etiology of Bell's palsy remains unclear although microcirculation failure of vasa nervorum , ischemic neuropathy , infection ,genetics and immunological causes

process include herpes simplex virus type 1 (HSV-1), herpes simplex virus type 2 (HSV-2), human herpesvirus, varicella zoster virus (VZV), influenza B, adenovirus, Coxsackie virus and Epstein–Barr virus (EBV)

The majority of patient will recover completely

poor prognosis has been related to :

complete paralysis at onset or incomplete paralysis with late onset of recovery, old age, a dry eye, abolished taste, absent stapedius reflex and postauricular pain.

Normal function is usually regained within three months in about two-thirds of all patients. No further recovery is expected after a period of six months has elapsed

Exercises

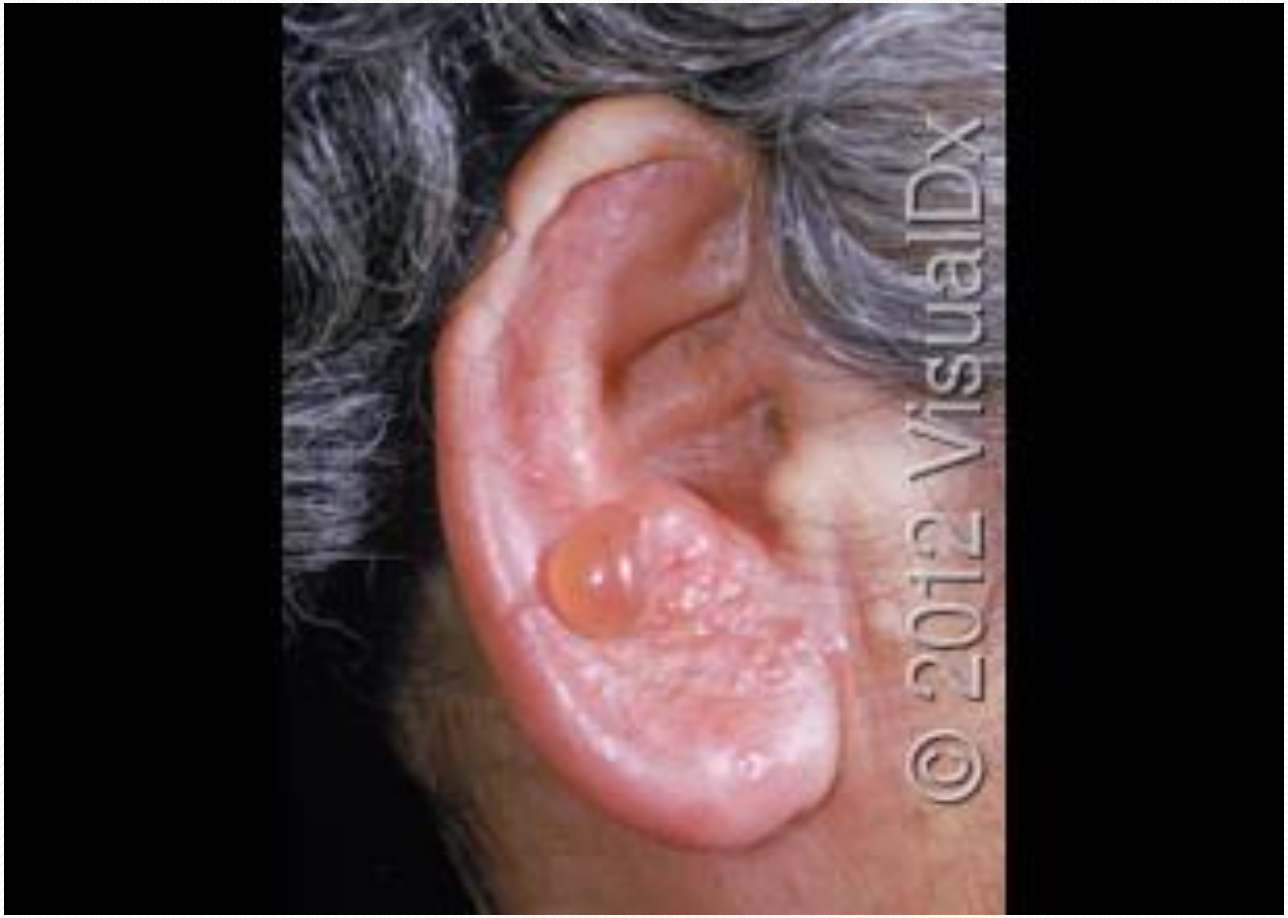
Predinsone 1mg /kg for 5 days then followed by ten days taper

Acyclovir 200-400 mg 5 times daily for 10 days

Facial nerve disorder of viral origin

VARICELLA ZOSTER VIRUS INFECTION (RAMSAY HUNT SYNDROME

Ramsay Hunt syndrome is a peripheral facial nerve palsy accompanied by an erythematous vesicular rash on the ear (zoster oticus) or in the mouth , The mechanism of disease is reactivation of the latent VZV virus in the **geniculate** ganglion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The onset of palsy is preceded by pain which may persist and be excruciating. In a small proportion of patients, the facial palsy is accompanied by a sensorineural hearing loss

The prognosis for Ramsay Hunt is worse than Bell's palsy. Persistent weakness is observed in 30–50 percent of patients and only 10 percent recover completely after complete loss of function without treatment

Treatment

Same as Bell's palsy (2 -3 weeks)

Predison 1 mg / kg / day for 5 days follwe by 10 days taper

IV acyclovir 250 mg 3 times daily or 800 mg orally 5 times daily

Facial nerve trauma

management of facial nerve paralysis following trauma is generally deferred until the patient is both medically and neurologically stable

MAXILLOFACIAL TRAUMA

stab wound or mandible fracture

Treatment by end to end anastmosis or interposition graft

TEMPORAL BONE TRAUMA

Longitudinal fracture---- 20 % facial palsy perigenigulate region

Transverse fractures ------ higher incidence of facial nerve paralysis (50 percent) and the labyrinthine and mastoid segments are most commonly involved

Middle ear and mastoid surgery

The most common site of injury during middle ear or mastoid surgery is the distal tympanic segment including the second genu, followed by the mastoid segment

If an injury to the facial nerve is recognized intraoperatively, exploration with decompression of proximal and distal segments of the nerve should be undertaken

If more than 50 percent of the circumference has been disrupted it should be repaired with either direct suture or graft

Parotid surgery

Cerebellopontine angle tumour surgery

NEONATAL FACIAL NERVE INJURY

Forceps delivery

more than 90 % good prognosis

Facial nerve paralysis as complications of the ear infection

- Otits media

Facial n paralysis may complicate both acute and csom due to due direct involvement of the nerve by infection through Fallopian canal dehiscence or through Fallopian canal osteitis with bone erosion and nerve involvement

- MALIGNANT OTITIS EXTERNA

Malignant otitis externa is an invasive Pseudomonas or Aspergillus infection of the ear canal which may lead to skull base osteomyelitis

Facial palsy indicates advancing infection and invasion through the bony-cartilaginous junction and the fissures of Santorini, under the tympanic ring and posteriorly to the stylomastoid foramen.

Primary or secondary

Primary facial nerve tumours are rare. Schwannomas and haemangiomas are the most frequent

Any part of the nerve may be involved and multiple segments can be affected simultaneously

Clinical features slowly progressive for n function , recurrent palsy and pain

Treatment

Poor facial n function -------- resection and graft

Good facial n function ------- conservative treatment with regular imagining and clinical evaluation

Secondary facial nerve tumors

Squamous cells carcinoma or adenoid cycstic carcinoma of the parotid gland

if the facial nerve is functioning preoperatively, the nerve can be preserved in most patients

The facial nerve should be sacrificed if there is direct invasion of the tumour into the nerve where the tumour cannot be separated

Neurophysiology of pain

pain as ‘an unpleasant sensory or emotional experience associated with actual or potential tissue damage

Most otalgia is mediated via unmyelinated pain fibres, which characteristically cause a diffuse dull ache. Myelinated fibres, such as supply skin or dental enamel, are associated with much better localization and easier diagnosis.

Pain may be nociceptive or neuropathic

or central nervous systems or from an abnormality in the pain processing system. The resulting sharp, sudden, stabbing, lightning type of pain is typical of neuralgias

Nerve supply of the ear

•1.The auriculotemporal branch of V innervates the anterosuperior external canal and pinna, but also the temporomandibular joint.

2. The facial nerve makes a smaller contribution, providing some sensory input from
the posterior tympanic membrane and external canal and the bowl of the concha.

3. Cranial nerve IX innervates the posterior external canal, meatus and tympanic membrane,
but also the ipsilateral oropharynx. Its tympanic branch (Jacobson's nerve) forms the tympanic plexus,
innervating the middle ear cleft.

4. The auricular branch of the vagus (Arnold's nerve) has a similar otologic distribution, but cranial nerve X has a vast dispersion to the viscera of the neck and even mediastinum

5. The upper cervical nerves C2 and C3, via the great auricular nerve and lesser occipital nerve, supply the cranial surface of the pinna, but also the skin and muscles of the neck and cervical spine.

This rich innervation of the ear allows central misinterpretation of the origin of

Causes

1.from the ear 2. Refereed

From the ear

- From the pinna
- Trauma : tear , laceration , bite
- Heamtaoma
- Infected eczema
- Perichondritis
- Infected basal or squamous cell carcinoma

b) from the meatus

1.impacted wax

2. impacted foreign body

3. otitis externa

4.Herps zoster oticus

5.keratosis obturance

6. furnculosis

7. malignant otitis externa

8. carcinoma

C) middle ear

1.bullous myringitis

2. traumatic perforation

3. OME

4.carcinoma

5. acute om

6. otitis baro trauma

7. hemotympanum

D)mastoid

1.acute mastoiditis

2. zygomatic mastoiditis

3. Bezold's abscess

4. complications of cholesteatoma

5. cholesterol granuloma

E) inner ear

1.noise

2. menieres disease

3. tinnitus

4. vestibular shwannoma

Causes of referred otalgia

- Via the V cranial N
- Lesion of the teeth and jaw

Impaction of molar tooth , apical abscess , dental caries , malocclusion , TMJ arthritis

- Lesion of salivary gland and duct (acute infection or calculus)
- Sphenopalatine neurolagia
- Lesion of the tongue , ulceration , carcinoma

B)via the X and IX CN

1. lesion of the oro and hypophayrnx

. acute phayrngitis and tonsillitis

.parapharyngeal and retropharyngeal abscess , quinsy

. tonsillectomy , TB, neoplasm

- 2.Lesion of the tongue

Ulceration , neoplasm , infection

3.elngated styloid process causing stretching of the glosspharyngeal CN

4. Glossphayrngeal neuralgia

C)via the 2nd and 3rd cervical spinal nerve

Cervical disc lesions

Arthritis of the cervical spine

Fibrositis of the upper part of steronmastoid m

The most common cause of refereed otalgia are impaction of lower molar tooth , infection or removal of tonsil , and dental malocclusion

How to arrive at diagnosis

_ **History**

– ***Features suggestive of primary otalgia (due to ear disease):***

_ hearing loss;

_ aural discharge;

_ vertigo;

_ unilateral rather than bilateral symptoms.

– ***Symptoms suggesting referred otalgia:***

_ pain on chewing/trismus;

_ dysphagia/odynophagia;

_ hoarseness;

_ risk factors (smoking/alcohol history);

_ neck swelling/goitre;

_ cervical musculoskeletal symptoms;

_ dental history/recent treatment.

– ***Features of neuropathic pain:***

_ radiation, e.g. to throat;

_ typical time course/duration;

_ quality of pain;

_ trigger zone/precipitating factors, e.g. swallowing.

_ **Examination**

– Primary otalgia:

_ inspection of ear and otoscopy;

_ palpation for tenderness;

_ aural examination with teleotoscope and microscope;

_ tympanometry.

– Referred otalgia:

_ cranial nerve (CN) examination, especially CN V, VII, IX and X;

_ palpation of cervical lymphatic chain;

_ assessment of cervical spine mobility/ tenderness;

_ palpation of TMJ and pterygoid muscles;

_ exclude trismus;

_ dental inspection for caries, absent dentition and malocclusion;

_ direct and fibreoptic examination of oropharynx and laryngopharynx;

_ palpation of oropharynx to seek induration, trigger zone or styloid bone

– Where diagnosis eludes the examiner, CT will detect skull base erosion, petrous apex disease and otherwise asymptomatic malignancies and demonstrate the styloid process. Enhanced MRI is superior in evaluating soft tissue disease, e.g. cranial nerve lesions, such as vestibular schwannoma or adenoid cystic carcinoma of the infratemporal fossa